

Late rapid deterioration after endoscopic third ventriculostomy: additional cases and review of the literature

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Object. Late rapid deterioration after endoscopic third ventriculostomy (ETV) is a rare complication. The authors previously reported three deaths from three centers. Three other deaths and a patient who experienced rapid deterioration have also been reported. Following the death at the University of Toronto of an additional patient who underwent surgery elsewhere, they canvassed pediatric neurosurgeons in North America, Europe, Australia, and Asia for additional cases.

Methods. An email was sent to the members of the Canadian Congress of Neurological Sciences who are pediatric neurosurgeons, to the pediatric neurosurgery email list of the American Association of Neurological Surgeons, to the email list of the International Society for Pediatric Neurosurgery, and to designated neurosurgeons in the United Kingdom, France, Japan, Korea, Taiwan, and Australia, who in turn contacted pediatric neurosurgeons in their countries. A data form was provided, and data from previously reported cases were extracted.

Nine additional cases were identified, and the results were collated with those of the seven cases previously reported. Patient age at surgery ranged from 2 days to 13 years (mean 7.6 years). The most common causes of hydrocephalus were aqueductal stenosis in 50% of patients and tectal glioma in 25% of patients. The time to treatment failure ranged from 5 weeks to 7.8 years (mean 2.5 years). Thirteen patients died, one patient was in a vegetative state, one patient was mildly disabled, and one patient whose condition deteriorated outside the operating room was alive and well. In the 13 patients in whom the ventriculostomy site was visualized at autopsy or repeated endoscopy, the ventriculostomy was shown to be occluded.

Conclusions. Late rapid deterioration is a rare but lethal complication of ETV. The mechanism is unclear, but deterioration can occur long after the ETV becomes occluded. Patients and caregivers should be counseled regarding this potential complication. An indwelling ventricular access device is an option for patients undergoing ETV.

KEY WORDS • endoscopic third ventriculostomy • adverse outcome • pediatric neurosurgery

ENDOSCOPIC third ventriculostomy has been used increasingly to treat hydrocephalus associated with obstructive hydrocephalus, both de novo and in patients who have indwelling shunts and who usually present with a shunt obstruction.^{3,5,15,23} Recently, the application of the procedure has been expanded to patients with hydrocephalus associated with fourth ventricular outlet obstruction,

Dandy–Walker malformation,¹⁹ Chiari malformation,^{2,6,17,21} and communicating hydrocephalus, including normal-pressure hydrocephalus.^{9,16} Although generally good results have been reported in most series, with success rates (defined as no further surgical procedures required) of approximately 70%,⁴ rare but serious complications have also been reported. They include vascular injury, hypothalamic injury, and occasionally death.^{10–12,20,22} Surgeons and most patients often prefer ETV to shunt implantation because of the anticipated long-term complications of CSF shunt insertion, which are well known.⁷ The acute complications of ETV and the potential for early failures have been well reported in the literature. We have also reported a series of three patients

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; ETV = endoscopic third ventriculostomy; EVD = external ventricular drain; GCS = Glasgow Coma Scale; ICP = intracranial pressure; MR = magnetic resonance; UK = United Kingdom.

demonstrating that the procedure can be associated with late rapid deterioration and death.¹⁰ The frequency of this problem has been uncertain. As is often the case in patients who experience adverse events, information is frequently not published, although additional cases have been noted anecdotally at international meetings. We made a concerted effort to identify as many additional cases as possible to obtain an idea of the frequency of this complication, to discern any common features that might explain its pathogenesis, and to identify risk factors so that it can be avoided. In this report we add an additional nine cases to the seven currently reported in the literature.

Clinical Material and Methods

An email was sent to the members of the Canadian Congress of Neurological Sciences who are pediatric neurosurgeons, to the pediatric neurosurgery email list of the American Association of Neurological Surgeons, to the email list of the International Society for Pediatric Neurosurgery, and to designated neurosurgeons in the UK, France, Japan, Korea, Taiwan, and Australia who in turn contacted pediatric neurosurgeons in their countries. A data form was provided, and it was requested that it be returned to the primary author. Data from previously reported cases were extracted.

Results

In Table 1, data pertaining to all of the patients are summarized, including those of previously reported cases. In Table 2, further details on the previously unreported cases are collated. The cause of the treatment failure was aqueductal stenosis in eight patients, tectal glioma in four patients, congenital hydrocephalus in three patients, and intraventricular hemorrhage in one patient. There were nine

male and seven female patients. Age at the initial surgery ranged from 2 days to 13 years (mean 7.6 years). The time from surgery to a sudden acute deterioration in condition ranged from 35 days to almost 8 years (mean 2.5 years). In 13 patients who underwent either a repeated endoscopy or in whom an autopsy was performed, the third ventriculostomy was occluded. Its status in the other three patients is unknown. Thirteen patients died, one patient was in a vegetative state, one patient was mildly disabled, and one patient, whose condition deteriorated outside the operating room just prior to surgery, was alive and well. In Table 2, further details extracted from questionnaires pertaining to the new cases are summarized. Most of the surgeons at the centers had a great deal of experience performing ETVs and followed up a large group of patients. There did not appear to be any unique features in how the ETVs were performed. Most surgeons used blunt perforation of the floor of the third ventricle, followed by balloon dilation of the opening. There were no intraoperative complications, and only one patient experienced them postoperatively. There was no reduction in the size of the ventricles in three patients, mild reduction in three patients, and moderate reduction in three patients (in one case [Case 12] the ventricles reexpanded to pre-ETV levels, although the patient was asymptomatic). The symptoms and signs at delayed presentation were almost universally headache, vomiting, and lethargy followed by obtundation. The family delayed in recognizing the problem in two cases, but the emergency department staff recognized it in all cases. Medical and surgical management consisted of urgent treatment of raised intraventricular pressure and an attempt to reestablish CSF drainage. Autopsies were performed in three patients. In the following section, we describe four previously unreported cases.

Illustrative Cases

Case 8

History and Operation. This 8-year-old girl presented with headache and papilledema. Magnetic resonance imaging demonstrated triventricular ventriculomegaly, periventricular edema, and a presumed tectal glioma (Fig. 1). The girl underwent an uneventful ETV at a center in another country and experienced resolution of her symptoms and the papilledema. Postoperative MR imaging showed resolution of the periventricular edema and a flow void (Fig. 2); cine MR imaging performed 14 months after surgery demonstrated the same results. The patient moved to the greater Toronto area and despite a referral to a pediatric neurosurgeon was not followed up.

New Presentation. Two years and 6 months after her first procedure, the patient presented to another hospital emergency department with a 2-week history of headache that the family attributed to a flulike illness. After she underwent CT scanning, which demonstrated ventricular enlargement, periventricular edema, and obliteration of the basal cisterns (Fig. 3), she suffered a rapid deterioration in her level of consciousness, experienced respiratory arrest, and had fixed, dilated pupils.

Treatment and Outcome. Intubation and mechanical ventilation were initiated, and the patient was given intravenous mannitol, and transferred quickly to our center. On arrival, she

TABLE 1

Summary of data pertaining to cases in which late rapid deterioration occurred after ETV*

Case No.	Country	Age (yrs), Sex	Indic ETV	Previous Shunt	FU (mos)	ETV Occluded	Outcome
1	UK	3.0, M	IVH	no	4.0	unknown	death
2	UK	11.0, M	AS	no	4.0	unknown	death
3	Australia	4.0, M	CH	no	84.0	yes	death
4	Canada	13.0, F	TG	no	42.0	yes	death
5	UK	0.8, M	AS	yes	7.0	unknown	death
6	Canada	0.3, M	AS	no	24.0	yes	death
7	France	13.0, F	TG	yes	24.0	yes	healthy
8	US	10.8, F	TG	no	30.0	yes	death
9	UK	13.6, F	TG	no	60.0	yes	death
10	UK	12.9, M	CH	yes	57.0	yes	death
11	France	3.3, M	AS	yes	9.0	yes	death
12	France	7.8, F	AS	no	94.0	yes	disability
13	UK	4.8, M	CH	no	1.4	yes	death
14	US	9.9, M	AS	no	5.0	yes	mild disability
15	US	5.8, F	AS	yes	1.2	yes	death
16	Japan	7.9, F	AS	no	22.0	yes	death

* Cases 1 through 7 were previously reported. Means for patient age and follow-up period were 7.6 years and 29.8 months, respectively. Abbreviations: AS = aqueductal stenosis; CH = congenital hydrocephalus; FU = follow up; indic = original indication for ETV; IVH = intraventricular hemorrhage; TG = tectal glioma.

TABLE 2
Further details of previously unreported cases*

Contributor Data		Acute Delayed Presentation										Autopsy Performed				
		Operative Data					Follow Up			Delay in Recognition Because of Absence of Shunt						
		Case No.	No. of ETV's Performed at Center	Year of ETV	Technique to Open 3rd Ventricle Floor	Complications†	Reduction of Ventricles After ETV	Flow Void Through Floor of Ventricle	Persistent Symptoms	Duration of Symptoms	Symptoms			Signs	By Family	By Health Provider
8	25	2001	laser wire (cold), FB	no	no	none	yes	no	14 days	headache, vomiting, then lethargy	obtundation	yes	no	intubation, hyper, mannitol	EVD	yes
9	200	1999	monopolar wire	no	mild	none	not done	no	1 day	headache	obtundation	no	no	intubation, hyper, mannitol	ventricular tap, EVD, repeated ETV	no
10	200	1998	figure-8 balloon	no	none	none	no	no		headache, vomiting	obtundation	no	no	found dead in bed	attempted re-suscitation only	yes
11	159	2003	forceps	no	moderate‡	not done	yes	no	2 hrs	lethargy, coma	obtundation	yes	no	intubation	none	no
12	250	1994	monopolar wire, figure-8 balloon	yes	moderate‡	yes	yes	no	6 hrs	headache, vomiting	obtundation	no	no	intubation, hyper	ventricular tap, CSF shunt, EVD, repeated ETV	no
13§	66	2001	laser fiber, FB	yes	moderate	not done	not done	yes	4 days	headache, vomiting	none	no	no	intubation	EVD, decompression of CM	yes
14	90	1999	blunt probe, FB	no	mild	yes	yes	no	2 days	headache, vomiting, lethargy, weakness	neurological deficit, hemiparesis	yes	no	none	CSF shunt	no
15	90	1996	blunt probe, FB	no	none	yes	yes	no	<1 day	headache, vomiting, lethargy	obtundation, hemiparesis	no	no	intubation	EVD	no
16	120	1995	FB	no	mild	yes	yes	no	1 day	headache, lethargy	none	no	no	intubation, hyper, mannitol	EVD, repeated EVD, CSF shunt	no

* CM = Chiari malformation; FB = Fogarty balloon; hyper = hyperventilation; OR = operating room.

† There were no complications during surgery. The only complications were postoperative.

‡ The reduction of the ventricles in this patient was moderate but then expanded again.

§ This complex case involved a history of renal failure, peritoneal dialysis, multiple episodes of peritonitis, a planned intraabdominal renal transplant when the patient was an adult, and immunosuppression. Surgeons wanted to avoid shunt insertion if possible.

|| This patient died after living for 2 years in a vegetative state.

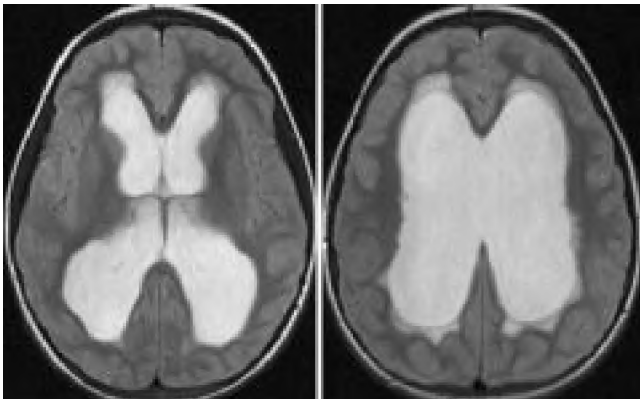


FIG. 1. Case 8. Axial T₂-weighted MR images obtained before the ETV showing ventricular enlargement and periventricular edema.

had fixed, dilated pupils and no brainstem reflexes. She underwent emergency needle aspiration of the right lateral ventricle followed by insertion of an external ventricular drain. She made no significant neurological recovery, and follow-up MR imaging demonstrated multiple areas of ischemia in the medulla oblongata, cerebellum, midbrain, thalamus, basal ganglia, hippocampus, and left occipital hemisphere (Fig. 4). Electroencephalography showed slow background activity and no reactivity. Death occurred rapidly following withdrawal of life support.

Pathological Findings. At autopsy, the third ventriculostomy was found to be occluded (Fig. 5) with scarring 1.2 mm thick, indicating that the occlusion had probably been present for months (Fig. 6). In addition to multiple areas of ischemia, there was a typical tectal glioma.

Case 9

History and Operation. This girl was born at 34 weeks' gestation at another hospital. There were no particular concerns during the neonatal period, but when she was 6 months of age it was noticed that her head circumference was larger than the 97th percentile. In 1991, a ventriculoperitoneal shunt was inserted at another neurosurgical center and the shunt became infected on two occasions.

In October 1999, the patient was admitted to the hospital at the age of 9 years after a 3-week history of intermittent headaches. By the time she was transferred to the neurosurgery center, she had a GCS score of 10 and a CT scan showed triventricular hydrocephalus. The patient was transferred to the operating room and an ETV was undertaken with the aid of a flexible scope. The hole in the floor of the third ventricle was made using the tip of a figure-eight balloon, which was then inflated to make the stoma wider. A temporary EVD was placed, and during the same session the old shunt system was removed. On cannulating the ventricle, the pressure was markedly raised.

Postoperative Course. Postoperatively, the patient's headaches resolved. On the 3rd postoperative day, the EVD was removed because a CT scan showed that the ventricles had decreased in size. The patient was discharged home on the 4th postoperative day and was subsequently followed up in the outpatient department. During a routine visit, it was noted that her left leg was smaller than her right, and an MR



FIG. 2. Case 8. Sagittal (A) and axial (B and C) post-ETV T₂-weighted MR images showing a flow void through the floor of third ventricle and resolution of the periventricular edema.

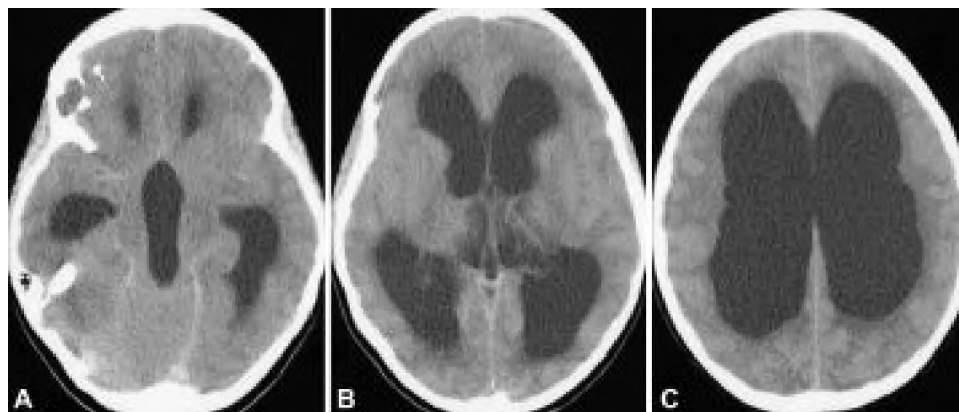


FIG. 3. Case 8. Unenhanced axial CT scans obtained when the patient presented following rapid deterioration 2.5 years after the first procedure indicating recurrence of the periventricular edema and obliteration of basal cisterns.

imaging study of her head and spine was undertaken. This study confirmed that the ventricle had remained smaller than it was on admission; somewhat surprisingly, however, it revealed an asymptomatic Chiari malformation Type I.

In addition to being assessed in the neurosurgical department, the patient's condition was assessed by staff in the orthopedic department, who noted a leg length discrepancy of 1 cm but found no scoliosis. Because the patient was completely asymptomatic from the perspective of the Chiari malformation Type I, she was merely monitored.

New Presentation. The patient remained well until July 2004, when she was 13 years of age. She experienced a sudden onset of headaches, which began at 4 p.m. This symptom was soon accompanied by vomiting, and the patient was taken to her local emergency department at 10 p.m.

Examination, Treatment, and Outcome. The patient's GCS score was 15 but it temporarily decreased to 8 after she was given codeine phosphate. Her GCS score returned to 14 and she underwent CT scanning, which confirmed triventricular hydrocephalus. Shortly after the scan was obtained, the patient became apneic and bradycardic (30 beats/minute). Both pupils were fixed, and her GCS score decreased to 3 at 12:20 a.m.

Intubation and mechanical ventilation were initiated, and the girl was transferred directly to the operating room at 2:15 a.m., where she was found to be hemodynamically unstable. Her pupils remained fixed and dilated despite administration of 100 ml of 20% mannitol. An emergency procedure was undertaken whereby a cannula was inserted into the ventricle and CSF was found to be under high pressure. An endoscope was also inserted to confirm that the floor of the third ventricle had no patent stoma. An EVD was left in situ but, unfortunately, there was no clinical improvement. The following day, brain death was declared.

Case 10

History and Operation. This 7-year-old boy presented with bilateral papilledema, increasing unsteadiness of gait over a 6-month period, and a recent poor performance at school. An MR image revealed triventricular hydrocephalus and a tectal plate lesion. There was no medical history of note, but his head circumference on admission was great-

er than the 98th percentile. The ETV was performed using a rigid endoscope. The hole in the floor of the third ventricle was made using a monopolar wire (no current) and was then enlarged using a figure-of-eight balloon. At the time of the operation, CSF was collected but was found negative for CSF markers. No EVD was left in situ; postoperatively the patient was well and was discharged home on the 4th postoperative day.

Follow-up imaging showed resolution of the papilledema and improvement in the ataxia; improvement in school performance was also noted. Subsequent MR imaging studies showed no significant reduction in ventricular size and no CSF flow across the floor of the third ventricle was seen on an MR imaging flow study. The tectal plate tumor remained unchanged. The last imaging study was obtained in January 2002. At that point, the patient was well, and al-

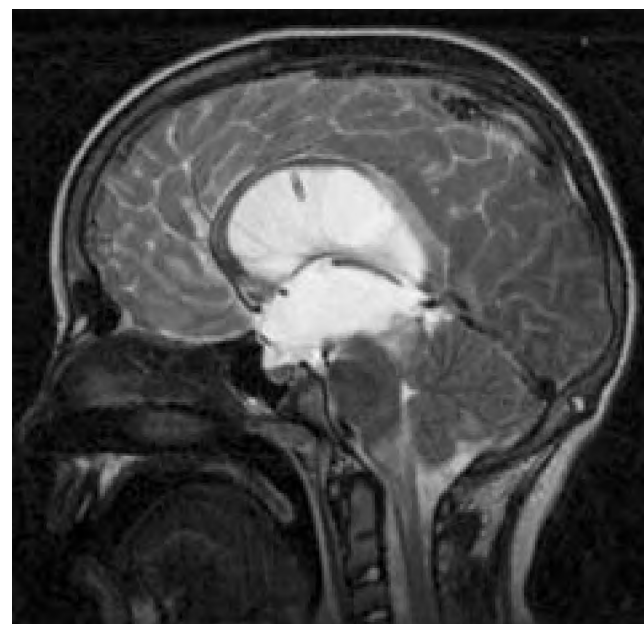


FIG. 4. Case 8. Sagittal T₁-weighted MR image after EVD insertion showing a tectal glioma and ischemic injury to the brainstem from tonsillar herniation.

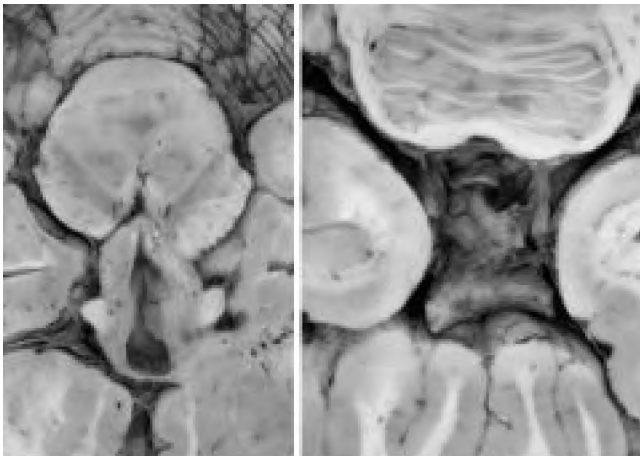


FIG. 5. Case 8. Postmortem photographs of the floor of the third ventricle from the exterior (*left*) and interior (*right*), revealing occlusion of the ETV.

though monitoring of ICP was considered, it was believed reasonable to manage the case using an expectant observation policy.

New Presentation and Outcome. The patient remained well with no headaches until July 2003 when he was on holiday with his family and began to complain of headaches at 10 p.m. Shortly after this, he started to vomit and both symptoms persisted until 4 a.m., when he appeared to fall asleep. His mother went to check on him at 6:30 a.m. and found him unconscious. He was transferred to the local hospital, where he was found to be asystolic and, despite resuscitation, was declared dead.

Pathological Findings. A postmortem examination confirmed that the patient died of acute hydrocephalus and that the third ventriculostomy site “appear[ed] to have been covered over by a thin membrane.” The postmortem examination also confirmed the tectal plate astrocytoma.

Case 12

History and Operation. In utero hydrocephalus was diagnosed in this patient at 38 weeks’ gestation based on ultrasonography findings. An in utero MR imaging study obtained at 39 weeks revealed the presence of triventricular hydrocephalus and aqueductal stenosis (Fig. 7A). This finding was confirmed on an MR image obtained on the 1st day of life (Fig. 7B), and the girl underwent an uneventful ETV the following day.

Postoperative Course. Subsequent MR images obtained at 8 days and at 2 months initially showed decompression of the ventricles (Fig. 7C); however, MR images obtained at 4 months and at 3.5 years showed reexpansion of the ventricles and loss of the flow void (Fig. 7D). Nonetheless, the patient remained asymptomatic. Her condition was seen to be unchanged on an MR imaging study obtained when she was 5.5 years of age, with similar expansion of the ventricles and loss of the flow void through the floor of the third ventricle. The patient played rugby and soccer, did very well in school, and had a full-scale intelligence quotient of 124 when tested at 5 years of age. Her neurological examinations continued to be normal.

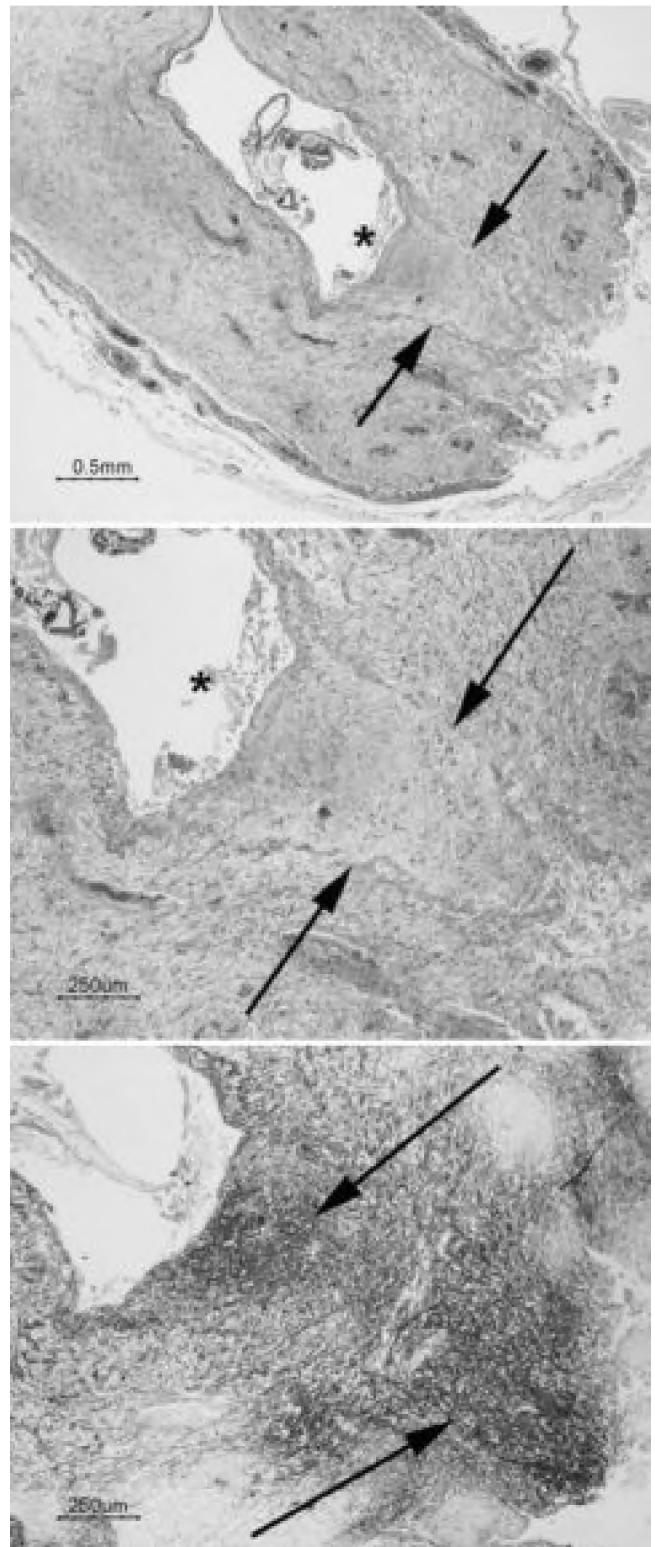


FIG. 6. Case 8. Photomicrographs of the floor of the third ventricle. (bottom right of images) showing occlusion and wedge-shaped scar tissue (*arrows*) approximately 1.2 mm thick. Asterisks mark the interior floor of third ventricle. On immunohistochemical analysis, the scarred area stained with glial fibrillary acidic protein (*lower panel: arrows*).

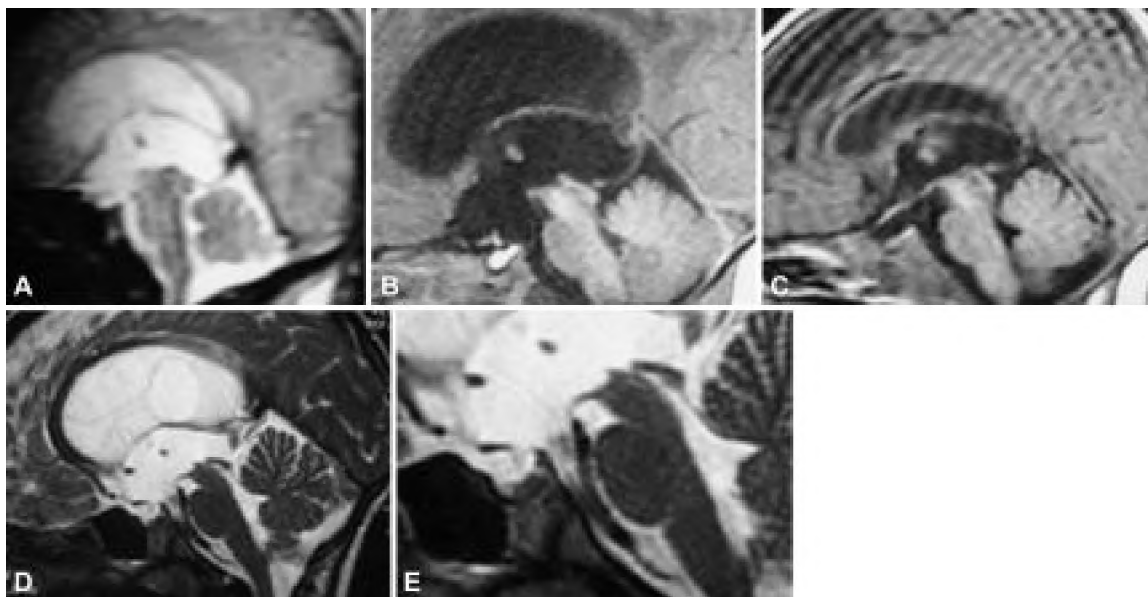


FIG. 7. Case 12. Magnetic resonance images. A: In utero image obtained at 39 weeks' gestation showing lateral and third ventricular enlargement caused by aqueductal stenosis. B: Sagittal T₁-weighted image obtained on the 1st day of life, showing lateral and third ventricular enlargement and aqueductal stenosis, along with downward ballooning of the third ventricular floor. C: Sagittal T₁-weighted image obtained 12 months after the ETV showing resolution of hydrocephalus, a decrease in the size of the lateral and third ventricles, and elevation of the floor of the third ventricle. D: Sagittal T₂-weighted image obtained 3.5 years post-ETV showing recurrence of the ventricular enlargement and downward ballooning of the floor of the third ventricle. The patient was asymptomatic at that point. E: Sagittal T₂-weighted image obtained 5.5 years post-ETV showing the ventricular size to be unchanged; the patient was still asymptomatic.

New Presentation. The patient remained well until she was almost 8 years of age; one day at 5:00 p.m. she began to complain of a slight headache. At 6:00 p.m., she began to vomit and by 11:00 p.m. she was unconscious. At 3:00 a.m., she was admitted to the neurosurgery department in that condition and with decerebrate posturing.

Treatment and Outcome. The patient underwent an immediate ventricular puncture, followed by insertion of a ventriculoperitoneal shunt. Because the improvement in her condition was slight, an EVD was inserted at 8:00 a.m. the following morning, and this procedure was followed by repeated ETV and removal of the EVD. Unfortunately, the girl made a minimal recovery and was left in a vegetative state.

Discussion

Late rapid deterioration after ETV is clearly a rare complication, but it is often fatal. That only 16 cases could be assembled from the literature and an email-based survey speaks to the rarity of this event, although it is difficult to exclude ascertainment bias. In addition, patients who experienced the event and who were salvaged may not have been reported. This uncertainty makes accurate estimates of the incidence impossible. Rough estimates based on the frequency of this complication as a total of the number of ETVs performed may be in the general range of 1 or 2 per 100 cases. In our own center in Toronto, Canada, one death occurred for approximately 150 ETVs; in Leeds, UK, two deaths occurred in approximately 200 procedures (P. Chumas, personal communication, 2005); in Paris, France,⁴ one patient in a vegetative state was reported in approximately

200 ETVs; and in Sidney, Australia,¹⁸ one death occurred in approximately 250 ETVs. Death following the complications of CSF shunt obstruction or infection is a much more familiar event to most neurosurgeons, particularly in patients whose conditions rapidly deteriorate often a long distance from an acute care setting. However, the number of patients with shunts is much greater. In a series of 907 patients with CSF shunts followed up over a 10-year period, 10 of 124 deaths (most others were due to neoplasms) were directly attributable to CSF shunt failure.²⁴ These data indicate that as uncertain as these estimates are, the death rate following ETV may not differ from that seen with CSF shunts. Accurate estimates of the incidence of rapid deterioration occurring in patients who have undergone either CSF shunt insertions or ETVs can be obtained only through outcome studies that are large, prospective, and long term.

Defining what constitutes late rapid deterioration following ETV is also somewhat arbitrary. Given the average treatment failure rate of approximately 30%, it is clear that many patients experience failures following ETV, many presumably with recurrence of presenting symptoms, failure of the ventricles to decrease in size, CSF leak, and so forth. What distinguishes the patients in this report in their symptomatology is that they died rapidly or almost died after an extended interval during which their symptoms had been relieved by an ETV. The definition of late is also somewhat arbitrary, and the categorization of the patient in Case 15, whose ETV failure occurred 35 days after the procedure, could be questioned in terms of this definition. In the mind of the particular surgeon, however, the patient had recovered from what appeared to be a successful ETV prior to experiencing sudden deterioration. A common theme is that the patients appear to be doing very well prior to the

event. When the patients present, it is with signs and symptoms of raised ICP, and imaging studies as well as operative findings confirm the recurrence of hydrocephalus. What is disturbing is how quickly the patients experience decompensation; despite rapid efforts to reestablish CSF drainage, the outcome is almost universally poor.

The pathogenesis of late rapid deterioration is not clearly known. With this expanded series of patients, it appears clear that the ETV becomes occluded. In several cases, the ETV was occluded for some time. In Case 8, a detailed autopsy was directed at the floor of the third ventricle in particular; the gliotic scarring over the ETV site was 1.2 mm thick, indicating that the floor had been occluded for several months. In Case 12, neuroimaging showed occlusion of the ventriculostomy based on the recurrence of downward ballooning of the floor of the third ventricle and loss of a flow void 3.5 years following the surgery; yet the child's condition did not rapidly deteriorate until 4 years later. Although the path of CSF absorption in patients suffering from aqueductal occlusion is unknown, transcerebral filtration and/or resorption by the choroid plexus seem plausible pathways. In instances of ventriculostomy occlusion, the same pathways are presumably reinitiated. It is possible that changes in brain compliance occur following successful ETV so that patients are more susceptible to any perturbations, as are patients with shunt-treated hydrocephalus. Recent evidence supporting the role of the nasal lymphatics in CSF absorption^{13,14} suggests that something as benign as an upper respiratory infection might impede CSF absorption enough to start the intractable rise in ICP. Demonstrated evidence of ventriculostomy occlusion occurring well before the onset of symptoms in some patients after an ETV raises the possibility that patients who have shunts might have a similar experience. Most neurosurgeons infer that the onset of symptoms in patients who have shunts occurs with complete occlusion of the system. It may be, however, that some patients tolerate an absence of flow for some time, through alternative pathways. This may explain how, in patients with documented asymptomatic shunt failure—due to increased ventricles or perhaps a disconnected shunt—symptoms of shunt obstruction can subsequently develop.

The failure of the patients, families, or emergency personnel to recognize the seriousness of the recurrence of symptoms of increased ICP are possible contributors to poor outcomes, although in the additional cases reported here, only two families failed to recognize the problem initially, and all emergency personnel appeared to react appropriately. Counseling patients and their families about “the internal CSF shunt” may improve patient awareness and compliance for follow up. Patients should undergo follow-up MR imaging studies that examine the patency of the ETV. At what point follow up can be discontinued is not known; it may depend on the cause of the hydrocephalus. Probably more important is that the patient and family understand that the “internal CSF shunt” could become blocked years after the operation.

All of the patients in whom an autopsy or repeated ETV was performed were found to have an occluded ETV. One presumes that there would also be absence of a flow void across the floor of the third ventricle on MR imaging, although there may not be an absolute correlation.⁸ The question arises as to what, if any, treatment patients should be

offered if no flow void is found or was never demonstrated. Certainly these patients need to be counseled about the possibility of acute deterioration. If there is loss of a flow void and reexpansion of the ventricles (as occurred in Case 12), a repeated ETV appears to be a reasonable option. If this is not technically possible or the patient declines the operation, then either a CSF shunt or CSF reservoir may be alternatives. Certainly these patients need to be continuously monitored.

Several authors have recommended inserting ventricular access devices at the time of ETV, to possibly save patients who experience sudden rapid deterioration.¹⁸ These devices have the added advantage of allowing monitoring of ICP via a needle puncture in patients having symptoms any time after surgery. Disadvantages are the potential for CSF leak or infection, which may be as high as 5%.¹ Although it is not possible to make a strong recommendation for or against ventricular access devices, certain patients (such as those living in remote locations) may benefit more from their implantation.

Conclusions

Late rapid deterioration is a rare but lethal complication of ETV. It appears to occur in typical candidates for this procedure around the world. It can be preceded by an absence of flow void on MR imaging, and complete occlusion of the ETV stoma was found in all of the patients who were examined at autopsy or endoscopically. Clinical deterioration can occur long after the ETV becomes occluded and can be very rapid. Patients and caregivers should be counseled regarding this potential complication and should not be left with the notion that the hydrocephalus is cured. An indwelling ventricular access device may be an option for providing rapid access to the CSF space in an emergency situation.

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